CASE REPORT

Dysmenorrhoea presentation in Herlyn-Werner-Wunderlich syndrome: A case study

Maryam Noor Malik, Gulshan Ara, Tasneem Akhtar, Humera Naeem

Abstract

Herlyn-Werner-Wunderlich syndrome is a rare congenital anomaly characterized by uterus didelphys, obstructed hemi-vagina, and ipsilateral renal agenesis. This study aims to present a case of Herlyn-Werner-Wunderlich syndrome seen at Shifa International Hospital in Islamabad, Pakistan, in March 2024.

A 32-year-old married woman who presented with dysmenorrhoea for two years and cyclic abdominal pain. Abdominal examination revealed a tender mass in the right iliac fossa. Ultrasound showed uterus didelphys, absent right kidney, a distended endometrial cavity with complex fluid, and a pelvic adnexal cyst. MRI suggested differentials including exophytic leiomyoma, adnexal cyst, or endometriosis. She was planned for hysterectomy and myomectomy. Intraoperatively, two separate uterine bodies with individual adnexa were identified, confirming Herlyn-Werner-Wunderlich syndrome. Both the uteri were preserved. The patient was discharged after two days and advised to undergo a hysterectomy at the age of 40.

Atypical presentation with dysmenorrhoea and abdominal pain can delay diagnosis, increasing risk of complications like infertility and endometriosis.

Keywords: Dysmenorrhoea; Müllerian ducts; Uterus.

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Introduction

Herlyn-Werner-Wünderlich (HWW) syndrome is a rare form of paramesonephric (Mullerian) duct anomalies. HWW is characterised by the triad of uterus didelphys, obstructed hemi-vagina, and absence of a kidney on the same side.[1] The actual prevalence varies from 0.1%-3%. The patient typically presents with an abdominal mass, dysmenorrhoea, and abdominal pain resulting from haematocolpos.[2] This syndrome was first identified by Purslow in 1922, in a young female who exhibited progressively worsening pelvic pain, regular menstruation, and a pelvic mass.[3] The study presents a rare case of a woman with uterus didelphys, dysmenorrhoea, and absence of right kidney, diagnosed through clinical presentation and findings.

Department of Gynaecology and Obstetrics, Shifa International Hospital, Islamabad, Pakistan.

Correspondence: Maryam Noor Malik. e-mail: maryamnoorzeeshan@gmail.com
ORCID ID: 0009-0005-2592-1222

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Case Report

A married woman aged 32 years, G_4 P_2 A_2 (Gravida 4, Para 2, Abortion 2), presented to the outpatient department of Shifa International Hospital in Islamabad, Pakistan, in March 2024 with dysmenorrhoea persisting for the past two years. She reported right lower abdominal pain that began with the onset of the menstrual cycle. The pain was localised, with no radiation, and was not accompanied by vomiting, fever, or any urinary issues. She had a regular menstrual cycle accompanied by cyclic abdominal pain and dysmenorrhoea. For this issue, she received analgesics from a nearby pharmacy, which provided only temporary relief of the symptoms. She had a history of two first-trimester miscarriages, a preterm birth with early neonatal death (ENND) due to preterm premature rupture of membranes (PPROM) and respiratory distress syndrome (RDS), and later delivered a healthy baby girl at 36 weeks via spontaneous vaginal delivery. On admission, she did not have fever and her vital signs were stable, though she presented with mild anaemia. Other physical examinations of the patient were normal.

Abdominal examination revealed a tender mass in the right iliac fossa. Per-rectal examination revealed a mass in the pouch of Douglas. The patient was admitted to the gynaecology department, where she was treated with analgesics, paracetamol, and Omeprazole to relieve her symptoms. Standard investigations were conducted (Table). The patient's complete blood count (CBC) was

Table: Laboratory investigations.

CBC	Values	Normal range
Haemoglobin (g/dl)	11.6	12.0–15.5
ESR mm/hr	14	0–20
WBC's (10^9/L)	10	4–11
Neutrophils	58.6%	40–75%
Lymphocytes	30%	20–45%
Monocytes	6%	2–10%
RBC's (10^{12}/L)	4.3	4.2–5.4
Platelets (10^9/L)	430	150–450

within the normal range (Table). Urine R/E and microscopic examination revealed no signs of infection. Abdominal and transvaginal ultrasound (TVUS) showed the absence of right kidney and an enlarged endometrial cavity containing complex fluid, respectively (Figure 1). A preliminary diagnosis of haematocolpos, haematometra, and right kidney agenesis was considered. MRI of the pelvis was conducted for additional assessment. The MRI results suggested uterus didelphys, exophytic leiomyoma (myometrium), adnexal cyst, and endometriosis. The right renal agenesis was present (Figure 2). The patient was planned for myomectomy. Upon surgery (open laparotomy) and exploration of the pelvic cavity, it was discovered that the patient had two uteruses, each with its own fallopian tube and ovary (Figure 3). Two endometrial cavities were noted, with the right-sided tube blocked and a rudimentary horn present. Based on her history of dysmenorrhoea, pelvic mass, preterm delivery, recurrent miscarriages, single congenital kidney, as well as the investigations and intraoperative findings, the definitive diagnosis was ascertained as the patient having a rare condition of Herlyn-Werner-Wünderlich syndrome. Both uteri were preserved during surgery. No complications were observed during or after the surgery. After two days, the patient was discharged. She was instructed to return for follow-up regarding her dysmenorrhoea or any other issues arising out of the surgical procedure that was performed, and advised to undergo a hysterectomy at the age of 40. Written informed consent was obtained from the patient for publication of this case study and any accompanying images.

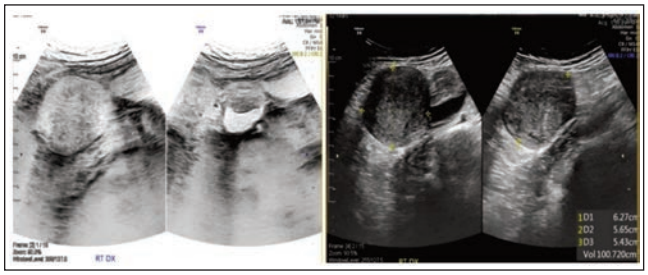

Figure-1: Ultrasound scan of the endometrium cavity with complex fluid.

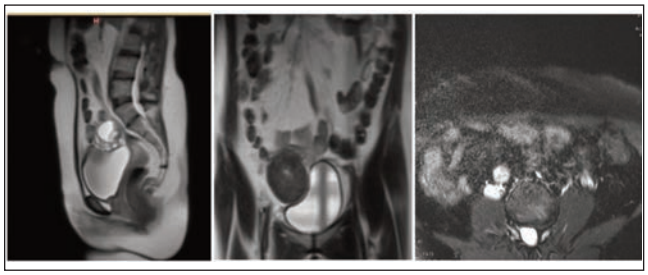

Figure-2: MRI scan right renal agenesis and adnexal cyst or endometriosis.

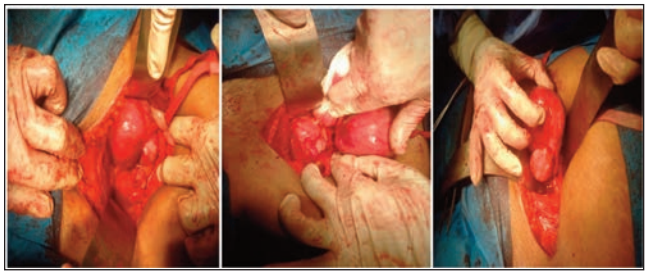

Figure-3: Uterus didelphys per op findings.

Discussion

The case describes Herlyn-Werner-Wüunderlich (HWW) syndrome in a young married woman. The MRI of the pelvis revealed the absence of the right kidney, with right endometrium cavity and cervix filled with blood. Based on the patient's history and clinical findings, most notably the pelvic mass and presence of a single congenital kidney, a diagnosis of HWW syndrome was established.

The literature indicates that cases may present with a variety of symptoms, including dysmenorrhoea, abnormal vaginal discharge, vaginal or pelvic mass, acute urinary retention, vomiting, fever, labour complications, infertility, endometriosis, or complicated pregnancy. In this case, although the diagnosis was delayed, once identified, appropriate corrective management was successfully implemented, resulting in full recovery and the restoration of regular menstruation without complications.

A triad presentation of HWW syndrome is characterised by uterine didelphys, obstructed hemi-vagina, and absence of one kidney.[2] The cause is unknown, but the development of embryology is affected by both environmental and genetic factors. In HWW syndrome, the disruption of both metanephros and paramesonephric systems occur.[4] The uterus, cervix, fallopian tubes, and upper 2/3 of vagina develop from ducts of paramesonephric. The duct originates from the urogenital ridge.[5] The ducts then run caudally, positioned laterally to ducts of mesonephric, and eventually meets and fuses with the ducts of paramesonephric from the opposite side in the midline to make cervix, uterus, and upper portion of the vagina.[5] When the ducts fail to fuse, they result in the formation of two hemi-uteri and hemi-cervices, leading to Müllerian anomalies.[6] Disruption of the metanephric diverticulum leads to one-sided absence of both ureter and kidney.[7] HWW syndrome is classified based on vaginal morphology into two main categories: Class 1 (complete hemi-vagina obstruction) and Class 2 (incomplete hemi-vagina obstruction), with each class having two subclasses.[8] The present case falls into subclass 1.1, characterised by two uteruses and a blind hemi-vagina.

Conclusion

The atypical presentation of non-specific abdominal pain

alongside regular menstruation makes the diagnosis of Herlyn-Werner-Wünderlich (HWW) syndrome particularly challenging, highlighting the need for heightened clinical vigilance. Early recognition depends on increased awareness of this rare anomaly. A multidisciplinary approach, integrating expertise from radiology, gynaecology, and paediatrics is crucial to prevent complications and achieve optimal outcomes.

Disclaimer: None.

Conflict of Interest: Gulshan Ara is the HOD of Obstetrics and Gynaecology at Shifa College of Medicine (SCM), Islamabad. She signed the HOD letter for the publication of this case report, In accordance with the journal's policy. She is also a co-author of this case report.

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Author Contribution:
MNM: Concept, design, preparation of draft, reviewing and agreement to be accountable for all aspects of the work.
GA: Concept, design, literature review and final approval.
TA: Concept, design, preparation of draft and reviewing.
HN: Concept and design.